Genital porokeratosis of mibelli

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Abstract

Porokeratosis of Mibelli is a disorder of epidermal proliferation in which many different clinical forms can be distinguished. Two male patients with a localized type of porokeratosis limited to the genitalia are reported. Later in life they developed an annular skin lesion with peripheral keratotic ridge. The histological examination of a biopsy specimen showed the characteristic features of porokeratosis. There was no family history of similar skin disorders and the patients were not on any drugs. Genital porokeratosis is probably underdiagnosed and we believe that these patients should be followed up on account of the precancerous potential of this disease.

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Introduction

Porokeratosis is a disorder of the keratinisation which presents with typical annular lesions varying in diameter between a few millimeters and some centimeters with a hyperkeratotic raised border. The centre is often atrophic. At present many clinical forms of porokeratosis are recognised: disseminated superficial porokeratosis, disseminated superficial actinic porokeratosis, palmoplantar porokeratosis, linear porokeratosis, giant porokeratosis and isolated lesions or plaque type porokeratosis of Mibelli.[1]

The disease is thought to be inherited as an autosomal dominant trait.[2] Sporadic cases later in life have been frequently linked to drugs such as thiazide diuretics and immunosuppressive treatments.[3 4] The aetiology is unknown. Fibroblasts underlying cornoid lamella present ultrastructural abnormalities which certainly contribute to the disorganization of collagen and to the epidermal changes typical of the disorder.[5] Cultured fibroblast studies showed genetical abnormalities with preferential involvement of the short arm of chromosome 3.[6] These chromosome abnormalities could explain the increased susceptibility to malignant disease in patients affected by porokeratosis.[6]

Case reports

Case 1

A 70 year old male was referred to our sexually transmitted disease service for the evaluation of a fixed drug eruption of the glans. On examination an annular lesion, 1 cm in diameter was also observed on his scrotum (fig 1). This lesion had a narrow raised hyperkeratotic border and a slight depressed centre. No other abnormalities were observed. The patient stated that this lesion had been present for some years and after an initial increase in size had remained constant. He also said there had been no symptoms and so he had never worried about it. Family history was unremarkable and the patient was not on drugs.

Case 2

A 40 year old male with a 5 year history of histopathologically confirmed isolated lichen planus of the nails was referred to our sexually transmitted disease service for the evaluation of an asymptomatic lesion on his penis, which had developed two years before and had increased slowly. Physical examination showed on the dorsal surface of the penis an annular lesion 1·5 cm in diameter with a narrow raised hyperkeratotic border and a central atrophy. The top of this border showed a furrow filled with keratin. The remainder of

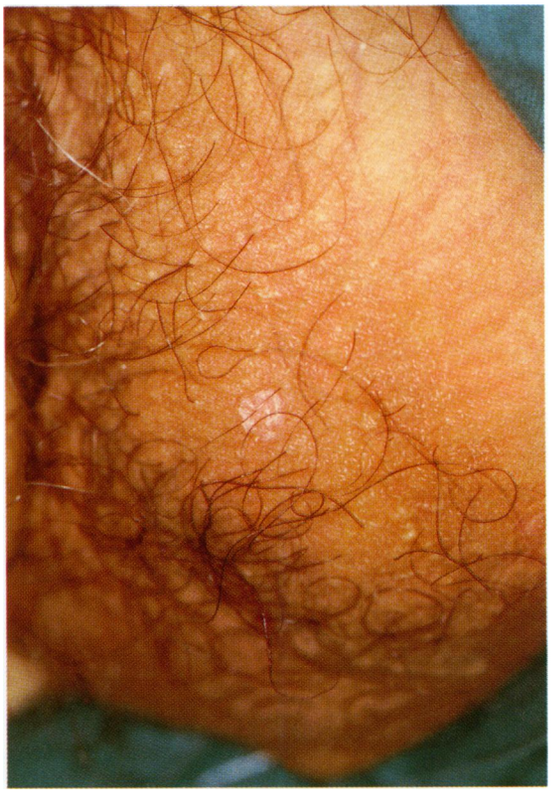

Figure 1 Annular lesion with narrow raised hyperkeratotic border on the scrotum.

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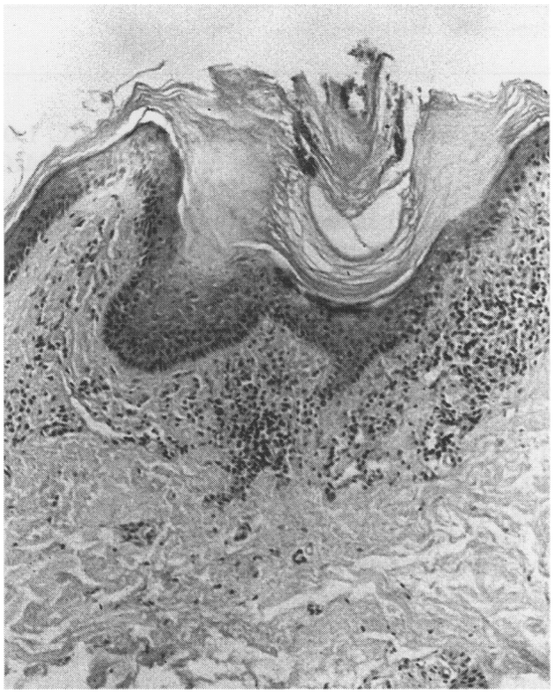

Figure 2 Histopathological examination: keratin-filled invagination of the epidermis with raised parakeratotic column (cornoid lamella).

physical examination only revealed 20 nail changes of lichen planus. There was no family history of similar skin disorders and the patient was not on any drugs.

Histopathological examination of a skin biopsy specimen of the border of the lesions showed, in both patients, keratin-filled invaginations of the epidermis with raised parakeratotic columns (so-called cornoid lamella) in the centre. The granular layer was missing beneath the cornoid lamella and single or clustered dyskeratotic cells and vacuolated keratinocytes were found at its base. In the stratum malpighii, some cells showed a premature keratinisation resulting in an eosinophilic cytoplasm. A non-specific perivascular infiltrate of chronic inflammatory cells was also present in the dermis (fig 2).

Discussion

Although porokeratosis of Mibelli (PM) can involve nearly any area of the body, the extensor surfaces of the extremities are the most common sites. Less frequently the mucosal surfaces are affected. Sometimes genital involvement is described in association with a generalised porokeratosis[7] while lesions limited to genitalia are considered very rare. The plaque type of PM actually appears most often on the limbs. Recently Levell *et al*[8] reported a 27 year old man with PM limited to the penis, scrotum and natal cleft. In this patient, as in our two cases, the disease is sporadic, developed later in life and did not appear to be linked to drugs,[3] immunosuppression,[4 9] hepatic or renal transplantations.[10]

PM with only one or a few lesions can be treated by cryotherapy, the CO_2 laser therapy[11] or by topical 5% 5-fluorouracil under occlusion[12] and good results with these techniques are reported in the literature. Both our patients declined any treatment because their lesions were asymptomatic and limited to a restricted area.

We think that the localisation on the genitalia of the plaque type of Mibelli is probably underdiagnosed. Both our patients actually consulted us for other skin disorders and the lesions were only discovered during physical examination without the patients mentioning them. Such cases should in fact be followed up since the association between malignant epithelial tumors and PM has been calculated to occur in 7%[13] to 11·6%[14] of cases. Moreover the localised type of PM in which the skin lesions develop later in life has shown a higher malignant transformation rate with a shorter latency period.[14]

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